Results: During the course of disease, 21 of 46 children with PME (45.65%) experienced total of 50 episodes of SE. The etiology was: neuronal-ceroid lipofuscinosis (7), Gaucher disease 2 (2), Nieman-Pick C (4), mitochondrial disorders (4), Lafora disease (2), Krabbe disease (1), KCNC1 (1). The age was 0.2-18 (mean 8.4) years. Nine patients experienced SE during the first year of disease, and in four cases, SE was the first epileptic event. All episodes had prominent motor symptoms: convulsive (25), myoclonic (13), focal including epilepsia partialis continua (12). Response to the treatment was variable, with common side effects. Most effective drug was midazolam in intravenous infusion with mean dosage of 0.35 mg/kg/h. The artificial ventilation was necessary in 7 episodes, in 4 together with circulatory support. Refractory SE was in 62% episodes, including nine SRSE. Recurrence rate was nearly 50%.

Conclusion: Children with PME frequently experience SE. Episodes are mostly convulsive, refractory to AEDs with high recurrence rate. SE appearance in later phase of disease contribute to prominent drug adverse effects. Managing SE in children with PME is challenge and requires rational approach in order to stop the seizure, and, on the other hand, to prevent side effects and worsening of general and neurological patient`s condition.

doi:10.1016/j.yebeh.2019.08.031

Epilepsy & Behavior 101 (2019) 106757

Refractory and super-refractory Status epilepticus- analysis of etiological factors

Cristina Munteanu, Stanislav Groppa
State University of Medicine and Pharmacy "Nicolae Testemiţanu", Chisinau, Moldova (the Republic of)
Institute of Emergency Medicine, Chisinau, Moldova (the Republic of)

Background: Status epilepticus (SE) can be a life-threatening condition associated with multiple complications, including death, and can progress to refractory and super-refractory SE. Treatment guidelines recently published partially addresses to the treatment of refractory and super-refractory SE.

Materials and Methods: We conducted a retrospective study of patients diagnosed as having SE- for a period of 2 years (01/01/2017-12/31/2018), according to the clinical presentation and EEG findings, also who had appropriate instrumental diagnosis included cerebral neuroimaging with CT and MRI to identify which condition are leading to refractoriness. The following data were analysed: age, sex, SE clinical type (convulsive and non-convulsive (NCSE)), neurological presentation, EEG features, etiology according to instrumental findings, neuroimaging study (brain CT and MRI), and/or blood examinations, and response to staged applied treatment protocol.

Results: Data are presented in raw numbers, data were analysed using Excel, student's t-test, we identified 78 patients, 29 of whom were female and 49 male, with a mean age of 42.2 $\pm$ 18.4 years. Regarding clinical SE type, convulsive SE was observed in 68 patients and non-convulsive in the remaining 10/78. As regards the SE type according to age patients with convulsive SE were older than patients with NCSE. According to EEG – focal SE was in 49 cases and generalised in 29, no difference for gender, for age focal SE more characteristic for older. Lesional SE was in 52 cases, toxic-dysmetabolic in 11 patients, anoxic SE in 3 and AED non- adherence in 12 from 78 patients. The oldest patients were those with lesional SE due to posttaumatic injuries, poststroke, brain metastasis, neurosurgical interventions followed by patients with non-adherence and toxico-dysmetabolic. Also it was observed among young patients the predominance of infectious and autoimmune underlying etiology (autoimmune/ viral encephalitis), and this cases proven to be refractory to first and second- line AEDs treatment (6/10 patients with refractory SE). 22 of patients have complete SE regression after being treated with benzodiazepines, 46 needed administration of second-line drugs like- phenytoin and phenobarbital and 10 patients – 12,8 % required anesthetic drugs to control the epileptic activity (3 patients- 3,8 % developed super-refractory SE (2 case herpetic encephalitis and 1 case anoxic brain injury).

Conclusion: Predictors of refractory status epilepticus were new diagnosis of SE and nonconvulsive SE. The etiology of refractory status epilepticus appears to be similar overall to that of nonrefractory status epilepticus, but more likely associated with encephalitis (viral encephalitis, in particular) and hipoxico-anoxic brain injuries. We also can conclude that good adherence to staged treatment as well as treatment of the underlying etiology is the key to success in controlling refractory seizures.

doi:10.1016/j.yebeh.2019.08.032

Epilepsy & Behavior 101 (2019) 106758

Long term follow-up of recurrent Status Epilepticus and Stroke-Like Episodes in a MELAS family

Lorenzo Muccioli, Francesca Bisulli, Lidia Di Vito, Chiara La Morgia, Laura Licchetta, Lara Alvisi, Valerio Carelli, Marco Zanello, Paolo Tinuper
IRCCS Bologna Institute of Neurological Sciences, Department of Biomedical and Neuromotor Sciences, University Of Bologna, Bologna, Italy

Background: Mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes (MELAS) is a disorder commonly caused by the A3243G/tRNALeu mutation of mitochondrial DNA (mtDNA). MELAS patients are at high risk of developing status epilepticus (SE) during stroke-like episodes (SLEs). We describe the long-term follow-up of 2 affected members of a MELAS family with recurrent focal SE associated with SLEs. SE treatment is discussed.

Method: A complete clinical work-out including clinical, biochemical, neuroradiological and EEG assessment was performed over the years in both patients.

Results: The mother developed since 33 years of age focal epilepsy (auditory and visual simptoms, hemiclonic seizures), sensorineural deafness, migraine, intestinal dysmotility, severe cognitive impairment, right homonymous hemianopsia and hemiparesis. She presented recurrent parieto-occipital SE associated with SLEs leading to death at 40 years of age.

Her son suffered photosensitive epilepsy since 17 years of age. He presented 7 episodes of occipital SE (elementary visual hallucinations and oculo-clonic seizures) associated with hemianopia, lactic acidosis and parietal-occipital SLE and refractory epilepsia partialis continua (EPC) in one occasion. SE became progressively more difficult to treat and complicated by lactic acidosis and rhabdomyolysis. During one SE propofol was used and the patient suffered multiple organ failure (propofol infusion syndrome-PRIS). Iv high-dosage midazolam was the most effective treatment of SE.

Both patients carried the mtDNA A3243G/tRNALeu mutation with a similar degree of heteroplasmy (80%).

Conclusion: We report the long term follow-up of 2 members belonging to a MELAS family with recurrent SE and SLEs. SE became refractory to treatment in both patients leading to death in the mother. Based on the occurrence of PRIS and evidence of mitochondrial toxicity,